The Surgical Treatment of Ventricular Septal Defect with Aortic Insufficiency, with Special Emphasis on Valve Replacement

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The surgical treatment of ventricular septal defect associated with aortic insufficiency was performed in 4 patients. Each case report was presented. The ventricular septal defect was closed first usually with buttressed mattress sutures without a patch in order to shorten the duration of myocardial anoxia. The aortic clamp was released intermittently with cardiac massage to resuscitate the heart. Simple closure of ventricular septal defect by itself improved aortic regurgitation in 2 cases. Resuscitation was impossible with cardiac massage and counter shock in 2 cases and a Starr-Edwards aortic prosthesis was implanted under intermittent or continuous coronary perfusion. One patient died in respiratory and renal insufficiency and the other survived. The authors discussed their present concepts regarding indication for operation and principle of operative procedure.

The surgical treatment of ventricular septal defect has become popular and many associated cardiac anomalies have been encountered. Among them, aortic insufficiency has aroused attention diagnostically to be differentiated from patent ductus arteriosus and therapeutically it has now been interested in the radical operation along with the development of prosthetic valve. Recently we experienced 4 cases of ventricular septal defect with aortic insufficiency and cured 3 of them by open-heart surgery. The purpose of this paper is to present our concepts regarding indication for operation and principle of operative procedure.

CASE REPORTS

Case 1. H. O., a 10-year-old boy, was found to have a heart murmur soon
after birth and had been cared not to do physical exercise after school age. He had had episodes of precordial pain especially during night since 2 months prior to admission and was referred to the University Hospital as a suspected case of patent ductus arteriosus.

Physical examination revealed an under-developed and under-nourished child (height: 124.7 cm., body weight: 22 Kg.). The precordium was protuberant and thrills were palpable. A grade 5/6 continuous murmur with systolic accentuation was best heard at the left sternal border in the third intercostal space and diastolic murmur at the apex (Fig. 1). Systolic murmur radiated well to the carotid artery which presented prominent impulse demonstrating Corrigan’s pulse. No cyanosis was noted. The blood pressure was 120/0 mm. Hg. Even after digitalization he had 3 episodes of paroxysmal dyspnea which was relieved by oxygen tent. At those times the recorded blood pressures ranged from 160/0 to 170/0 mm.Hg. Roentgenograms of the chest showed marked cardiac enlargement, predominantly of the left ventricle, with increased vascularity of the lung fields (Fig. 2). The cardio-thoracic ratio was 71 per cent. The electrocardiogram revealed marked left ventricular hypertrophy (Fig. 3).

Hemoglobin was 12.4 Gm./100 ml., erythrocyte count 4.59 million per cu. mm., hematocrit 37.5 per cent, CRP negative and ASLO under 100 Todd units. Nasal culture revealed normal flora.

Cardiac catheterization showed a left-to-right shunt in the right ventricle without hypertension in the right heart (Table I). Retrograde aortography showed regurgitation of dye into the left ventricle (Fig. 4). Although dye was not clearly seen in the right ventricle, it was definitely observed in slight degree in the pulmonary artery.

Operation was performed utilizing a median sternotomy, under the preopera-
diagnostic diagnosis of ventricular septal defect with aortic insufficiency. The left ventricle and pulmonary artery were markedly enlarged, but the aorta was scarcely dilated. Thrills were palpable over the right ventricular outflow tract just beneath the pulmonic valvular ring, and digital exploration of the interior of the right ventricle disclosed a dilated aortic cusp and jet stream beneath it, and the preoperative diagnosis was confirmed.

Extracorporeal circulation was established utilizing a rotating disc oxygenator, and total bypass was started when esophageal temperature reached 30°C. With the ascending aorta crossclamped, a transverse ventriculotomy was made.

### Table I. Cardiac Catheterization Data

|                  | Case 1 | | Case 2 | | Case 3 | |
|------------------|--------|--------|--------|--------|--------|
|                  | Pressure (mm. Hg) | O₂ Content (Vol. %) | pressure (mm. Hg) | O₂ Content (Vol. %) |
|                  | Mean | Syst./Diast. | | Mean | Syst./Diast. | |
| Pulmonary capillary | 7 | 30/19 | 14.50 | 8 |
| Pulmonary artery   | 22 | 30/0 | 15.90 | 20 | 38/10 | 15.40 |
| Right ventricle    | 15 | 11.60 | 12.65 | 18 | 40/0 | 17.85 |
| Right atrium       | 1.5 | | | 1 | 14.90 | |
| Superior vena cava | | | | | | 13.35 |
| O₂ capacity        | 20.00 Vol. % | | 20.05 Vol. % |
| Arterial O₂ content| 16.90 Vol. % | | 18.55 Vol. % |
| Arterial O₂ saturation | 84.5 % | | 92.6 % |
| O₂ consumption     | 225 ml./min. | | 190 ml./min. |
The ventricular septal defect was found just beneath the pulmonic valvular ring (Type I defect), which was large enough to pass through an index finger. The right coronary cusp prolapsed into the defect. The ventricular septal defect was closed with the reinforced 5 mattress stitches tied down over Teflon pledgets, and the aortic clamp was released gradually. The left ventricle was dilated markedly and cardiac resuscitation was impossible with cardiac massage and counter shocks. Accordingly, the aorta was clamped again and a transverse aortotomy was made. A Starr-Edwards aortic prosthesis (8-A) was sutured using horizontal mattress sutures under intermittent coronary perfusion after excising all aortic leaflets. The cardiopulmonary bypass lasted 106 min at a flow of 1,600 ml. per min. with moderate systemic hypothermia (the lowest rectal temperature 28°C and esophageal 29.5°C).

Postoperative course: Reoperation became necessary 11 hours later for hemorrhage which reached 900 ml. by midnight, at which time a bleeding point from an infundibular branch of the right coronary artery was sutured. However, respiratory resistance and râles in bilateral lung field had become marked since the original operation, and anuria was followed on the first postoperative day with gradual increase of serum NPN, BUN and potassium. The urinary output increased on the 4th and 5th postoperative day, but the respiratory insufficiency persisted. Cardiac arrests had occurred 6 times since the 3rd postoperative day and cardiac resuscitation was successful with external cardiac massage and counter shock at each time. However, he became gradually unconscious and died with cardiac arrest on the 5th postoperative day.

An autopsy showed that sutured parts of the ventricular septal defect and prosthetic valve were perfectly corrected and no other cardiac anomalies were present. The lung showed congestion and atelectasis which were mostly not due to bronchial secretory obstruction. The histology of kidneys showed the features of acute tubular necrosis with hemoglobin casts in renal tubules. Cardiac failure was considered to be the cause of death.

Case 2. M. A., a 14-year-old boy, had been under-developed and started to walk at 4 years of age when a heart murmur was noted. He was advised not to do
physical exercise owing to palpitation with exercise.

Physical examination: Height was 149.5 cm. and body weight 33 Kg. No cyanosis was noted. The precordium was protuberant, and thrills were most palpable at the left sternal border in the 2nd intercostal space (Fig. 5). A grade 5/6 continuous murmur with systolic accentuation was heard at the same place and the systolic murmur radiated to the neck where prominent pulsation was visible. The blood pressure was 140/0 mm.Hg.

The chest roentgenogram showed left ventricular enlargement with increased vascularity of the lung field. The cardio-thoracic ratio was 61 per cent (Fig. 6). The electrocardiogram revealed marked left ventricular hypertrophy (Fig. 7). Hemoglobin was 13.3 Gm./100 ml. erythrocyte count 4.64 million per cu.mm., hematorcit 40.5 per cent and CRP negative.

Retrograde acrtography demonstrated aortic regurgitation and confirmed the diagnosis of ventricular septal defect with aortic insufficiency (Fig. 8).

Operation was performed as in Case 1. The operative findings were almost same in the position and size of ventricular septal defect and the state of prolapsed aortic leaflet. The ventricular septal defect was sutured with 5 mattress sutures with Teflon pledgets. Cardiac resuscitation could not be obtained after release of aortic occlusion. Accordingly, an aortotomy was made, and a coronary perfusion cannula was inserted into the left coronary ostium for continuous perfusion. A Starr-Edwards aortic prosthesis (9A) was sutured excising the aortic valve including the right coronary cusp which was thickened and prolapsed into the left ventricle below other cusps. The cardiopulmonary bypass lasted 90 min. at a flow of 2,000
ml. per min. with moderate systemic hypothermia (the lowest rectal temperature 27°C and esophageal 29°C).

Postoperatively, respiration was assisted with a Benett respirator through a tracheostomy for 24 hours. The postoperative course was uneventful and he was discharged on the thirty-eighth postoperative day. The blood pressure was improved to 122/64 mm.Hg and the chest roentgenogram is shown in Fig. 9.

Case 3. T. K., a 10-year-old boy, was found to have a heart murmur at 2 or 3 months of age when he had a cold, but grew and developed normally without any symptoms, though he sweated often at night.

Fig. 6. Chest roentgenogram of Case 2.
Fig. 7. Electrocardiogram of Case 2.
Fig. 8. Retrograde aortogram of Case 2.
Fig. 9. Chest roentgenogram of Case 2 at the time of discharge. A prosthetic aortic valve visible.
Physical examination: Height was 131.7 cm. and body weight 26.7 Kg. No cyanosis was noted. There was a grade 5/6 systolic and grade 3/6 diastolic murmur, accompanied by a thrill, maximal at the left sternal border in the third intercostal space. The diastolic murmur was weaker than that in Cases 1 and 2. The blood pressure was 130/40 mm.Hg. The chest roentgenogram showed the cardio-thoracic ratio of 54 per cent and slightly increased vascularity of the lung field (Fig. 10). The electrocardiogram revealed marked left ventricular hypertrophy (Fig. 11). Cardiac catheterization showed a left-to-right shunt in the right ventricle without pulmonary hypertension (Table I). There was regurgitation of dye into the left ventricle in retrograde aortography (Fig. 12), but milder than in Case 1 and 2.

Fig. 10. Chest roentgenogram of Case 3.

Fig. 11. Electrocardiogram of Case 3.

Fig. 12. Retrograde aortogram of Case 3.

Fig. 13. Phonocardiograms recorded in Case 3. (A "to-and-fro" murmur).
In operation, the aorta and pulmonary artery trunk were isodiametric, and thrills were palpable over the right ventricular outflow tract just beneath the pulmonic valve, and intracardiac digital exploration determined the diagnosis. After a transverse ventriculotomy was made, a 1.5 × 1.0 cm. defect of Type I was found to be partly occluded by a prolapsed coronary cusp. The defect was closed utilizing two buttresses of Teflon, one of which was passed through beforehand with two armed sutures in order to shorten aortic occlusion time as possible. A vigorous cardiac beat returned with gradual release of the aortic clamp and cardiac massage, and a prosthetic valvular replacement was not necessary. The postoperative blood pressure was improved to 116/62 mm.Hg and the diastolic murmur became faint.

Case 4. K. O., a 12-year-old boy, has been sensitive to colds and was found to have a heart murmur at 4 years of age. There were no symptoms of diminished cardiac reserve except palpitation with exercise. One year before admission, he was first seen at the University Hospital Out Patient Clinic. There was a grade 4/6 holosystolic murmur, accompanied by a thrill, maximal at the left sternal border in the third intercostal space. No diastolic murmur was noted at that time.

Physical examination revealed a well-developed child (height: 140 cm., body weight: 31 Kg.) without cyanosis. Similar systolic murmur was present, but grade 3/6 early diastolic murmur had appeared in the aortic area and along the left sternal border (Fig. 13). The blood pressure was 112/48 mm.Hg.

The chest roentgenogram showed the cardio-thoracic ratio of 54 per cent and moderately increased vascularity of the lung field (Fig. 14). The electrocardiogram revealed marked left ventricular hypertrophy (Fig. 15). Retrograde aortography showed slight regurgitation of dye into the left ventricle (Fig. 16).

The operative findings were almost similar to Case 3. Thrills were palpable over the right ventricular outflow tract just beneath the pulmonic valve. A transverse ventriculotomy was made and a defect sized 1.5 cm. in diameter of Type I was found to be almost occluded by a prolapsed coronary cusp. The defect was
closed under intermittent aortic occlusion, utilizing a Teflon patch sewn to the defect edge by interrupted mattress suture, each reinforced with a Teflon pledget.

The postoperative course was uneventful and the blood pressure became 110/72 mm.Hg.

**DISCUSSION**

The incidence of association of aortic insufficiency in ventricular septal defect is reported to be almost 5 per cent. The 4 patients comprising the present series were observed during a period in which 74 patients with ventricular septal defect were operated at this University Hospital (Table II). Thus, the incidence of the association is about 5.4 per cent.

This association was described early by Laubry et Pezzi\(^1\) and Laubry,

Table II. Associated Cardiac Anomalies with Ventricular Septal Defect (1966. 8. 5)

<table>
<thead>
<tr>
<th>Condition</th>
<th>Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pure VSD</td>
<td>60</td>
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<tr>
<td>VSD+AI</td>
<td>4</td>
</tr>
<tr>
<td>VSD+ASD</td>
<td>3</td>
</tr>
<tr>
<td>Aneurysm of membranous ventricular septum</td>
<td>3</td>
</tr>
<tr>
<td>VSD+PS</td>
<td>1</td>
</tr>
<tr>
<td>VSD+ASD+PPS</td>
<td>1</td>
</tr>
<tr>
<td>VSD+LSVC</td>
<td>1</td>
</tr>
<tr>
<td>Multiple VSD</td>
<td>1</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>74</strong></td>
</tr>
</tbody>
</table>
Routier et Soulie. Since around 1950, the difficulty inherent in the differential diagnosis to patent ductus arteriosus has been emphasized. However, more recently the diagnosis has become not difficult with angiography if it is suspected.

The ventricular septal defect associated with aortic insufficiency is mostly situated immediately beneath the pulmonic valvular ring. The size ranges usually from 1 to 2 cm. in diameter. According to Dubost's review of 62 patients, the incidence of prolapsus of each aortic cusp is: right coronary cusp 51 per cent, non coronary cusp 20.8 per cent, right and non coronary cusp 9.7 per cent, left coronary cusp 1.6 per cent and undetermined 16.3 per cent, and a right coronary cusp is most frequently affected. Intracardiac digital exploration in a beating heart reveals an aneurysmally dilated affected aortic cusp herniated into the right ventricle partly occluding the septal defect with a relatively narrow jet flow around it. The affected aortic cusp is thickened, deformed and prolapsed into the left ventricle. Perforation of sinus of Valsalva rarely occurs. Calcification of the cusp is very rare. The ascending aorta is not so dilated as in acquired aortic insufficiency, however, the dilatation and hypertrophy of left ventricle are marked. Infundibular pulmonary stenosis is known to be accompanied, and 9 of the 18 cases in Keck's series had infundibular pulmonary stenosis. It was recognized in our patients that a right coronary cusp was prolapsed in Cases 1 and 2, however, none had infundibular pulmonary stenosis.

In catheterization of the right side of the heart, severe pulmonary hypertension is rare and pulmonary systolic pressure is mostly under 50 mmHg. The magnitude of the left-to-right shunt is relatively small. In 30 cases reported by Keck and Halloran, Y/Qpa under 29 per cent were 4 cases, 30 to 39 per cent 7 cases, 40 to 49 per cent 7 cases, 50 to 59 per cent 5 cases and 60 to 69 per cent 3 cases. In our patients, Y/Qpa was around 50 per cent, though arterial oxygen saturation was low and the data were somewhat inaccurate. The electrocardiogram shows marked left ventricular hypertrophy which is consistent not with the hemodynamic degree of ventricular septal defect, but with aortic insufficiency.

The development of aortic insufficiency is usually recognized with the appearance of a diastolic murmur along left sternal border in the follow-up observation of ventricular septal defect. Ash described already in 1950 that a case of ventricular septal defect who had been observed since 3 months of age suddenly fell into cardiac failure, and the pure systolic murmur up to that time had changed to a continuous murmur. Starr reported the development of aortic insufficiency at the age of 6 and 8 in his 2 operated cases. Later Spencer reported its occurrence at the age of 4, 5 and 6 in his 3 operated cases,
and Hollan described that aortic insufficiency occurred from $2\frac{1}{2}$ to 9 years of age at an average age of 6 years in 8 of the 12 observed cases. In our patients, the development of aortic insufficiency was at the age of 11 in Case 4 and not clear in others. Review of other reports and our experience indicate that most of aortic insufficiency associated with ventricular septal defect occurs not congenitally, but after birth.

The surgical correction of this combination of defects has been tried with closure of ventricular septal defect and reconstruction of aortic valve, but the results were not always satisfactory. Robinson indicated that closure of ventricular septal defect by itself might improve or recover aortic regurgitation in a relatively mild case. Asano, one of the authors has already pointed out this fact. Konno stated that severity of aortic insufficiency could be graded into three on the basis of retrograde aortographic findings, and patients in grade 3 needed repair of aortic valve, though simple closure of ventricular septal defect might improve or recover the insufficiency in patients in grades 1 and 2. He presented a case without recurrence of diastolic murmur even 6 years after simple closure of ventricular septal defect.

Simple closure of ventricular septal defect may not be suitable in instances with dynamically more severe regurgitation, and even cardiac resuscitation is often unsuccessful. However, the preoperative determination about the surgical procedure is sometimes difficult in relatively severe cases. Therefore, severe cases should be operated under the preparation of aortic valve replacement. Ellis and associates reported 19 surgical cases in which simple closures of ventricular septal defect, cusp advancement procedures and procedures designed to shorten the leading edge of the cusp usually resulted in persistence or recurrence of aortic valvular incompetence. They found that the most effective means was a prosthetic cusp replacement. However, the long term result of various prosthetic leaflet valves is discouraging in acquired heart diseases, and at present a Starr-Edwards ball valve or analogous valves are used mostly.

However, the application of a ball valve is handicapped by the facts that it is used in growing children, and the aortic orifice and ascending aorta are not so dilated in this disease. There may be no problem in the case in which a 9-A ball valve is inserted, but the case in which a ball valve of 8-A or smaller size may have trouble in future. Therefore, such an operation in severe cases should be deferred until the age of 12 to 14 years as Ellis stated. Case 1 might have problems in future, however, the operation was considered to be necessary, because the patient had angina and the prognosis was assumed to be poor in near future.

Operative procedure: As closure of the septal defect by itself may lessen the regurgitation, the right ventricle should be first opened. Garamella ad-
vocated the transaortic correction, but it seems to be difficult. The septal de-
fect must be closed with the ascending aorta crossclamped, and so the time re-
quired for the repair of the defect should be as short as possible. The ridge of
the ventricular septal defect is usually far from the conduction mechanism.
Accordingly, the defects is closed tight with buttressed mattress sutures without
a patch as mentioned before. Although the ventricular septal defects in our
patients were about 1 x 1.5 cm. in size, the defects could be closed perfectly by
direct suture in all instances except in Case 4 whose defect was sited relatively
low and apparently near to the conduction mechanism and so closed with a
patch.

The aortic clamp should be released intermittently with cardiac massage
to resuscitate the heart. If resuscitation is impossible, the ascending aorta is
crossclamped again and a prosthetic valve replacement is performed. Coronary
perfusion is necessary for the myocardium which had already anoxic arrest.
In Case 1, intermittent coronary perfusion with cold blood was utilized, never-
theless, cardiac resuscitation was delayed. On the other hand in Case 2, con-
tinuous left coronary perfusion was performed at a flow of 200 ml. per min.
for 57 min., and the heart was electrically defibrillated promptly with a single
AC counter shock (200 volt, 0.15 sec.). A continuous coronary perfusion can-
nula was designed, which can be made easily with a polyethylene tube (Fig. 17).
This was inserted into a coronary ostium and held in place by a suture applied
inside. This did not disturb the operative field and seemed to be very useful.
At the time of aortotomy closure, all air is evacuated from within the heart and
ascending aorta. The coronary perfusion cannula is withdrawn as late as
possible. The ventriculotomy wound is closed lastly.

Fig. 17. A coronary perfusion cannula designed by us. The tip is made of
polyethoeplyne.
SUMMARY

The surgical treatment in 4 patients with ventricular septal defect associated with aortic insufficiency is described. The closure of ventricular septal defect by itself improved the degree of aortic regurgitation in 2 moderately severe cases. However, aortic insufficiency could not be controlled by the closure in 2 severe cases, and aortic valve replacement was performed with a Starr-Edwards aortic prosthesis with one success. Our concepts regarding indication for operation and principle of operative procedure were presented.

REFERENCES